A RARE CASE OF MULLERIAN ANOMALY- (HEMATOMETRA WITH CERVICAL AGENESIS)

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ABSTRACT

Cervical Agenesis is a relatively infrequent mullerian anomaly. Sixteen years old, unmarried girl presented to ELMC&H Lucknow, with complaints of primary amenorrhoea and cyclical abdominal pain for 1 year. On Per-rectal examination, tense cystic mass was felt on right side, approximately 5x5 cm in size, tender and freely mobile. Cervix could not be palpated. Small knob like structure was felt on left side, 1.5x1.5 cm in size, firm in consistency and freely mobile. USG showed uterus of size 5x4x3 cm with collection in endrometrial cavity with? hypoplastic cervix suggestive of- Hematometra with cervical agenesis. On laparotomy, tense unicornuate uterus on right side and solid rudimentary horn on left side was found, which was attached to the uterus by peritoneal fold. Cervix was absent. Both ovaries and tubes were normal. Right cornua of uterus was completely excised leaving both the tubes and ovaries.

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INTRODUCTION

Congenital absence of the cervix is a rare condition and occurs 1 in 80,000 to 100,000 births. It is known to be associated with both partial and complete vaginal aplasia and may be associated with renal anomalies. According to the American Fertility Society, cervical agenesis is classified as a class IIb uterovaginal anomaly. Clinical presentation is usually with primary amenorrhea and cyclical lower abdominal pain, as was seen in our patient. Endometriosis or pelvic infection may result from the chronic hematometra by repeated manipulations.

CASE REPORT

A 16-year-old unmarried girl reported to gynae OPD with complaints of primary amenorrhea and cyclical abdominal pain for 1 year. There was no significant medical or surgical history. On examination secondary sexual characters were well developed. Abdomen was soft, non-tender, no organomegaly, On local examination, External genitalia was normal and pubic hair well developed. Per vaginal examination was not done .On per-rectal examination, tense cystic mass was felt on right side, approximately 5x5 cm in size, tender and freely mobile. Cervix could not be palpated. Small knob like structure was felt on left side, 1.5x1.5 cm in size, firm in consistency and freely mobile. All routine investigations and IVP were normal. USG showed uterus of size 5x4x3 cm with collection in endrometrial cavity with hypoplastic cervix suggestive of - Hematometra with cervical agenesis(figure-1). Examination was done under anesthesia. On per vaginum examination, lower 1/4 of vagina was well developed,rest of the findings were same. Patient was given an option of conservative surgery and counseled regarding the probable complications of the same and the need for hysterectomy later on. Patient and her attendants opted for direct hysterectomy because of prolong sufferings of the patient. Patient was taken for laparotomy. Per operative finding were right sided tense unicornuate uterus 5x4x3 cm with left sided solid rudimentary horn 1.5x1.5 cm, attached to uterus by peritoneal fold and cervix was absent. Both ovaries and tubes were normal (figure-2). Right cornua of uterus was completely excised leaving both the tubes and ovaries (figure-3) On cut section – uterus was filled with dark brown coloured blood around 25cc.(figure-4) Histopathology report showed Gross-Uterus(4.5x4x2cm) without cervix and adnexa. Microscopy-endometrial cavity showing evidence of haematometra, endometrium was nonsecretory.No cervical tissue was found. Post operative period was uneventful. Patient was discharged on 9th post operative day with the advise to come for vaginoplasty three months before marrige.

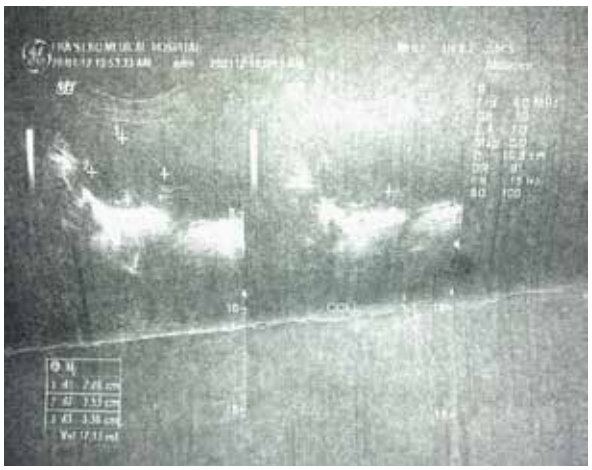

Figure1- Ultrasonography

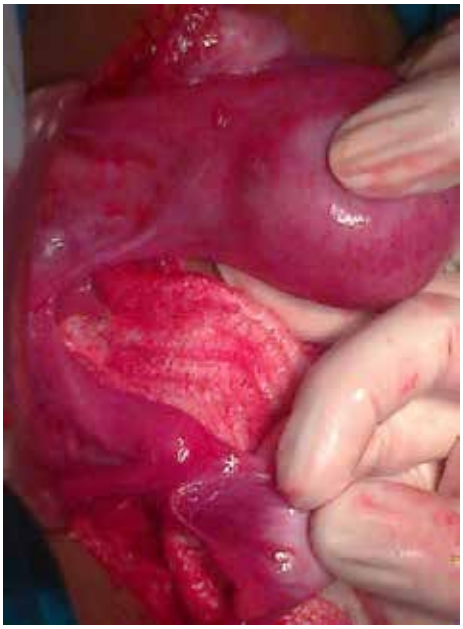

Figure2 *- Laprotomy finding*

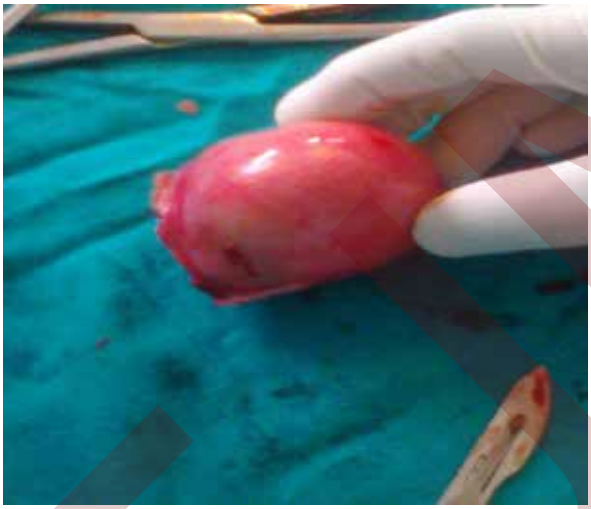

Figure 3 *- Gross Specimen*

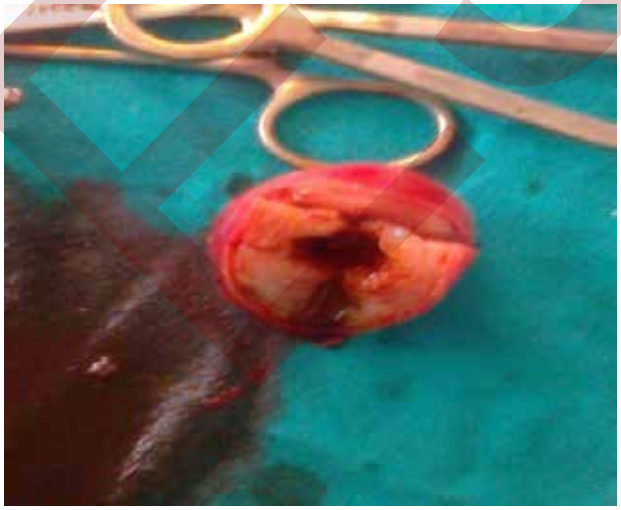

***Figure* 4** - Cut section

DISCUSSION

Patients with mullerian agenesis have a normal 46 XX karyotype, normal female phenotype, normal ovarian function. Typically, patients with mullerian agenesis present in adolescence with primary amenorrhea. Differential diagnosis of vaginal agenesis includes androgen insensitivity, low lying transverse septum and imperforate hymen. To manage vaginal agenesis effectively in young women, correct diagnosis of the underlying condition is important. Evaluation for associated renal (e.g. renal agenesis, pelvic kidney, duplex ureter) and skeletal anomalies are also essential [1, 2]

Mullerian abnormalities causing obstruction of blood discharge due to cervical or vaginal atresia are rare examples of malformation of the female genitalia. In the past, total hysterectomy represented a virtually systematic treatment of this type of condition. More recently, however, some attempts at preserving the uterus of these patients have been made in order to preserve their reproductive capacity[3]. This was achieved by creating a 'fistula' between the uterus and the vagina or between the uterus and a surgically formed neovagina, permitting normal flow between the endometrial cavity and the outer environment. These conservative techniques lead to relief of the pain due to blood accumulation, and provides the psychological benefit represented by menstruation and the possibility of maternity. Previously the recommended treatment for cervical agenesis was a hysterectomy because complications of renalizing the cervix were common and the possibility of a viable pregnancy was unlikely [4, 5]. Recent advances in reproductive technology and laparoscopic surgical techniques mean that conservative surgery is a possibility and perhaps should be considered as the first-line treatment option[6]. However the results of reconstructive surgery are better in patients with a well formed cervical body, with at least a palpable cord or only distal obstruction.

CONCLUSION

In patients with cervical agenesis, conservative laparoscopic surgery should be considered as first-line treatment. This surgery should be performed only in a highly specialized unit with the required expertise in laparoscopic surgery and in management of complex müllerian anomalies. Due to over all poor results of reconstruction for congenital absence of both the cervix and vagina, clinical experience suggest that conservative procedures can be worthwhile for a few carefully selected cases with adequate stroma to allow a cervicovaginal anastomosis.

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